

Long-term survival after univentricular heart surgery

Sunnegårdh J. (1), Synnergren M. (1), Dalén M. (2)

The Queen Silvia Children's Hospital, Sahlgrenska University Hospital, Institution of Clinical Sciences, Gothenburg University, Sweden (1); Department of Molecular Medicine and Surgery, Karolinska Institutet, Department of Cardiac Surgery, Karolinska University Hospital, Stockholm Sweden (2)

Objective. To examine long-term survival after surgery for univentricular heart defects.

Methods. All children operated due to univentricular heart defects in our institution before the age of 18 years from Jan 1st 1994 to May 31st 2017 were included. Patient files were analyzed and cross-checked as of May 31st 2017 against the National Population Registry allowing for reliable and complete data on survival. Primary outcome was all-cause mortality. Additionally, risk-factors of death were analyzed.

Results. 431 patients (41% female), median age and weight at first surgery of 10 days (quartile [Q]1, Q3: 6, 21) and 2.6 kg (Q1, Q3: 3.1, 4.1) were included. Mean follow-up time was 10.1 ± 8.9 years, with loss to follow-up of one patient. 108 deaths (25%) occurred with a median age at death of 88 days. Right ventricular dominance was found in 228 (52.9%) patients and left or biventricular dominance in 199 (46.2%), with a survival at 10 years of age of 63.3% and 89.9%, respectively ($p < 0.001$). No mortality difference between these two groups was found in conditional survival in patients surviving the first two years after surgery. In multivariate analysis, risk factors for death were severe AV-valve regurgitation before first stage palliation (hazard ratio [HR] 2.21, 95% confidence interval [CI] 1.00-4.85, $p = 0.049$), hypoplastic left heart syndrome (HR 1.79, 95% CI 1.10-2.92, $p = 0.019$) and right ventricular dominance (HR 2.32, 95% CI 1.31-4.10, $p = 0.004$). No significant difference was found in mortality at three years of age between patients born 1994-2004 ($n = 57/200$, 28.5%) as compared to those born 2005-2017 ($n = 40/182$, 22%). Heart transplantation was performed in 20 patients (4.6%) with four late deaths. As of May 31st 2017 34 patients were awaiting a stage 2 or 3 operation and 251 were alive with a complete Fontan circulation.

Conclusion. This study gives prognostic information of survival for patients undergoing surgical palliation due to univentricular heart defects. Patients with a systemic ventricle of left ventricular dominance have a significantly better long-term survival compared with patients with right ventricular dominance. However, this was due to a significantly higher mortality only during the first two years after the initial surgery.